Esophageal sarcoidosis presenting as pseudodiverticulum


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Abstract. Sarcoidosis is a multisystem granulomatous disorder that may involve many organs. However, the involvement of the gastrointestinal tract is very rare. This report describes an unusual case of esophageal sarcoidosis presenting as a pseudodiverticulum and reviews the world literature. Our case is also characterized by unusual progression of the esophageal involvement despite stable disease in other organs involved. Myotomy improved the pharyngoesophageal stenosis with no recurrence to date. Physicians should be aware of this rare manifestation, which should be suspected in any sarcoidosis patient complaining about dysphagia. (Sarcoidosis Vas Diffuse Lung Dis 2008; 25: 64-67)

Key words: dysphagia, non-caseating epithelioid granuloma, Zenker diverticulum

Case report

A 57-yr-old male consulted our hospital with a 18-month history of blurred vision of the right eye in June 1999. He had been previously diagnosed as having uveitis. Chest radiograph showed mild bilateral hilar lymphadenopathy and mild reticulonodular shadows (Fig. 1). Bronchoalveolar lavage fluid

Fig. 1. Chest radiograph showing mild bilateral hilar lymphadenopathy and ill-defined reticulonodular shadows.
(BALF) analysis revealed lymphocytosis (67%) and an increased CD4/CD8 ratio of 3.8 (upper limit of normal 3.5). Serum angiotensin-converting enzyme (ACE) activity was increased to 64 IU/L (normal < 40 IU/L). Pulmonary function tests and blood gas analysis were normal. The patient was asymptomatic. On the basis of these findings, he was clinically diagnosed as having pulmonary sarcoidosis (stage II) complicated with ocular involvement. Systemic steroid therapy (prednisolone 20 mg daily) was administered for ocular involvement. Because of the subsequent stable clinical course as evaluated by ophthalmoscopy, chest radiographs, pulmonary function tests and blood gas analyses, prednisolone was gradually tapered and stopped in 2003.

In March 2004, however, he became symptomatic with dysphagia when eating solid food. Chest radiographs, pulmonary function tests and blood gas analyses revealed no change. Chest computed tomography (CT) demonstrated enlarged pretracheal lymph nodes without tracheal compression, unchanged with the previous CT examinations. There was no adenopathy in the area of the upper esophagus. No pharyngeal involvement was seen on CT. In esophagography and a swallowing study, no complications were detected except mild gastroesophageal reflux revealed by esophageal pH monitoring. Serum ACE activity was slightly increased to 89 IU/L.

In June 2006, the reexamined esophagography suggested the possibility of a Zenker diverticulum in the upper esophagus (Fig. 2). Left-lateral cervicotomy with complete exploration of the hypopharynx and upper esophagus, however, revealed no signs of a cervical diverticulum. A complete lateral myotomy of the hypopharynx and proximal esophagus with a biopsy of the esophageal wall was performed. Histopathological examination showed non-caseating epithelioid granuloma in the wall of the esophagus (Fig. 3). Subsequently, the dysphagia for solid food improved. To date, the patient remained asymptomatic without the need for systemic steroid therapy.

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Fig. 2. Esophagography showing a diverticulum at the posterior wall of the upper esophagus. No esophageal stenosis is observed.

Fig. 3. Hematoxylin–eosin stain of the wall of the esophagus showing an infiltration consisting of multiple epithelioid cell granulomas with giant cells and some lymphocytes (20 x) (top), at higher magnification, classical non-caseating epithelioid granuloma with giant cells, infiltrating smooth muscle bundles (400 x) (bottom).
Here we report a case of esophageal sarcoidosis presenting with dysphagia, a pseudodiverticulum and severe pharyngoesophageal stenosis. Histopathology showed infiltration of the smooth muscle of the esophageal wall by sarcoid granulomas. There was no evidence of indirect compression of the esophagus by enlarged mediastinal lymph nodes.

Previous reports have presented various patho-genetic pathways for dysphagia in patients with sarcoidosis; 1) compression of the esophagus from outside the walls by enlarged lymph nodes (2), 2) achalasia associated with a contractile disorder of the cardia derived from neural invasion with granulomas and 3) direct infiltration of the esophageal wall by granulomas (3). Esophagogram, esophageal motility study and histopathological examination of the tissue obtained by esophagogastroscopy, thoracoscopy, mediastinoscopy or thoracotomy have been reported to be beneficial for the diagnosis of esophageal sarcoidosis (1, 4).

In summary, we have presented a case of pharyngoesophageal sarcoidosis demonstrating a pseudodiverticulum and pharyngoesophageal stenosis. Physicians should be aware of the possibility of this rare manifestation as cause of dysphagia in a patient with sarcoidosis.

**References**